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Congenital coronary artery fistula
Analysis of 17 cases

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A co-operative retrospective study of 17 patients with congenital coronary artery fistula was undertaken.

The right coronary artery was the most common vessel involved and the right ventricle was the site of drainage in the majority of the patients. Three methods of drainage of the fistula into the chamber are described. The majority of the patients were under 20 years of age and were asymptomatic. All the patients above the age of 35 were symptomatic. The symptoms and the cardiomegaly on the chest x-ray were well correlated with the size and duration of the shunt.

The physical signs, electrocardiograms, chest x-rays, cardiac catheterization data, and angiocardiographic features are described. Points helpful in the clinical diagnosis of this lesion are emphasized. Some aspects of the natural history of this lesion are described and indications for operation are mentioned.

A congenital communication of a coronary artery with a cardiac chamber or the pulmonary trunk is a rare but interesting lesion, for several reasons. It enters into the differential diagnosis of a continuous murmur which is a common physical sign in both the paediatric and adult cardiac patient. The anomaly may be the cause of congestive heart failure (Berman *et al.*, 1965; Honey, 1964) and may also present as a case of bacterial endocarditis (Sanger, Taylor, and Robicsek, 1959; Serratto and Kezdi, 1965). With the modern methods of cardiac catheterization and angiocardiographic examination, this lesion can be diagnosed accurately and is amenable to cure by the newer surgical techniques.

This report is based on the study of 17 patients. Six of them were not operated on and were followed for different times, thus providing an opportunity to document some aspects of the natural history. This paper also deals with the pathological anatomy, clinical features, and diagnosis, and discusses the indications for operation.

Patients and methods

For the purpose of this study we defined congenital coronary artery fistula as 'an abnormal communication between the coronary arteries or one or more of their branches into a chamber of the heart, or the pulmonary artery'. Excluded from this study were congenital communications of the coronary arteries with a chamber of the heart in association with atresia of the pulmonary or the aortic valve and intact ventricular septum, and patients with anomalous origin of the coronary artery from the pulmonary artery.

The diagnosis of the abnormal communication was made either by cardiac catheterization and angiocardiography or at operation.

A retrospective co-operative study of congenital coronary artery fistula was undertaken and 17 patients were admitted from various centres in the Netherlands and Belgium. One of us reviewed all the case histories of these patients and 2 or more of us studied in detail the serial electrocardiograms, chest *x*-rays, phonocardiograms, cardiac catheterization data, and angiocardiograms.

Results

Pathological anatomy The fistula originated in 10 cases from the right coronary artery and in 7 from the left.

The main right coronary artery was the most common site of origin (Table 1); this is in agreement with the published report of Neufeld *et al.* (1961).

The fistula was found to terminate most commonly in the right ventricle (9 cases), then in the right atrium (4 cases), the pulmonary artery (2 cases), the left atrium (1

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TABLE 1 *Congenital coronary artery fistula*

Origin of fistula		*No.*
Right coronary artery		10
Main right coronary artery	7	
Additional branch	2	
Circumflex branch	1	
Left coronary artery		7
Main left coronary artery	2	
Circumflex branch	2	
Anterior descending branch	1	
Additional branch from anterior descending	1	
Branches from circumflex and anterior descending	1	
Total number		17

TABLE 2 *Congenital coronary artery fistula*

Site of drainage		*No.*
Right ventricle		9
Right coronary artery	7	
Left coronary artery	2	
Right atrium		4
Right coronary artery	2	
Left coronary artery	2	
Pulmonary artery		2
Right coronary artery	1	
Left coronary artery	1	
Left atrium		1
Left coronary artery	1	
Both ventricles		1
Left coronary artery	1	
Total number		17

case), and both ventricles (1 case) (Table 2).

The affected artery was much dilated and tortuous. There were three types of drainage of the fistula (Fig. 1): (1) in 12 patients the involved artery entered directly into the chamber; (2) in 3 patients the involved artery terminated at the apex of the right ventricle in an aneurysm, which in turn opened into the chamber; and (3) in 2 other patients the affected artery ended in a convolution of small vessels, entering into the chamber through multiple openings. Only two patients had associated congenital cardiac lesions. One was a patient with tetralogy of Fallot in whom the fistula arose from the left coronary artery and ended in the pulmonary artery. The other patient had an associated persistent ductus arteriosus.

Clinical features The age of the patients ranged from 1½ to 65 years, but the majority were under 20 years of age (Table 3). The lesion was distributed equally in both sexes. There were 8 male and 9 female patients.

Presenting features Of the 17 patients, 12 were asymptomatic: these were referred because of the murmur with a diagnosis of persistent ductus arteriosus. In 2 instances the patient had been directly sent to the surgeons for ligation of the ductus. At operation no ductus arteriosus was found and further investigation revealed the fistula.

FIG. 1 *Angiocardiograms of the three types of drainage.* Left: *direct drainage of the left coronary artery* (arrow) *into the right atrium.* Centre: *filling of an aneurysm which in turn drains into the right ventricle through a small opening; a 'jet' is visible* (arrow). Right: *at the apex a convolution of a smaller vessel is formed* (arrow).

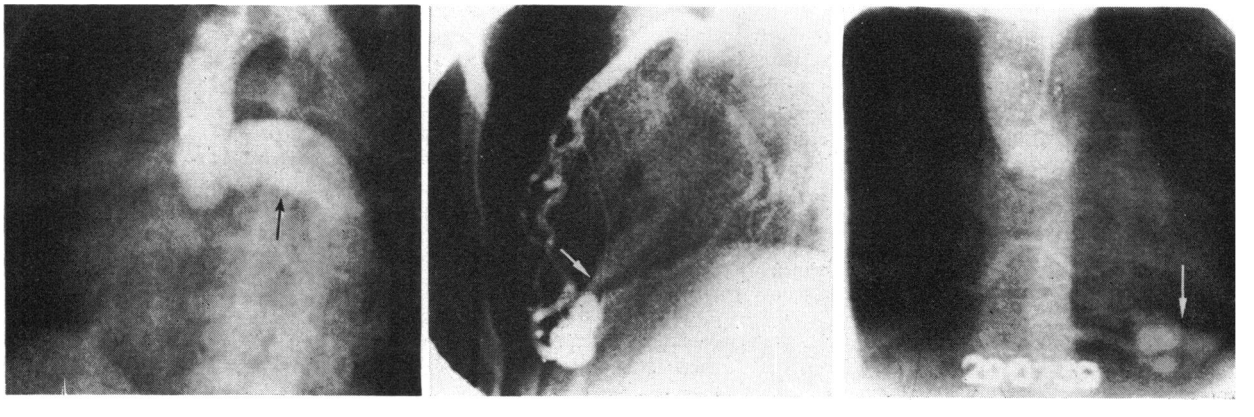


TABLE 3 *Congenital coronary artery fistula*

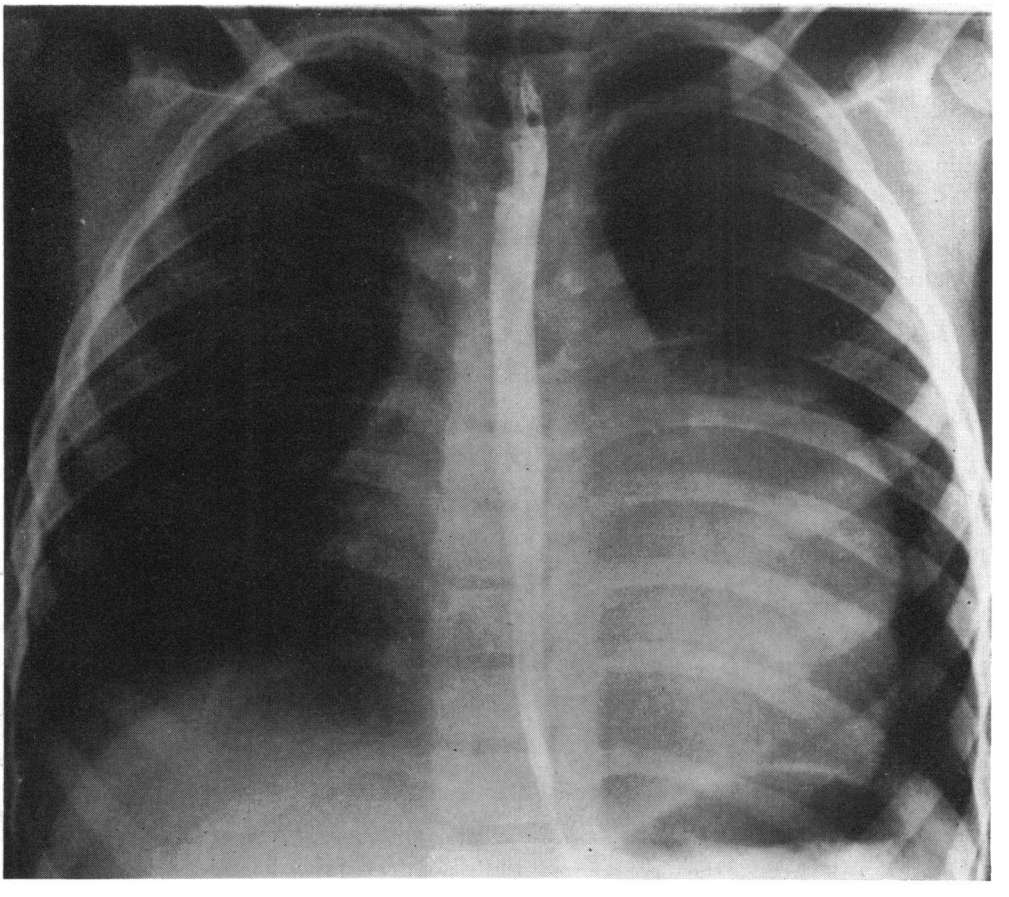
Age range in 17 cases: 1½–65 yr	
Age (yr.)	*No.*
0–9	8
10–19	5
20–29	1
30–39	1
40–49	1
50–59	—
60–69	1
Total number	17

The presenting features seem to be related to the size and duration of the shunt. Of the 14 patients under the age of 37, only 2 were symptomatic. One was the patient with tetralogy of Fallot; the other was a 6-year-old boy who was easily fatigued. The shunt in this case was more than 50 per cent of the pulmonary blood flow. In contrast, of only 3 patients over the age of 37, all were symptomatic: 2 patients, 47 and 65 years respectively, presented in congestive heart failure; the third patient presented with vague praecordial pain at the age of 37 and the electrocardiogram showed evidence of anterolateral infarction.

Physical examination The pulse pressure was well correlated with the size of the shunt. The larger the shunt, the higher the pulse pressure, except in one patient who had a very small shunt with a pulse pressure of 55 mmHg. A systolic thrill was present in 4 patients, in 3 of whom the fistula entered into the right atrium and in the other into the left atrium. None of the patients with entry of the fistula into either ventricle had a thrill, but Gasul *et al.* (1960) and Neufeld *et al.* (1961) have reported a thrill in patients with right ventricular communication of the fistula.

A continuous murmur was present in all

FIG. 2 *Chest x-ray of a patient with drainage of the left coronary artery into the left atrium. There is considerable dilatation of the left atrium, and normal pulmonary vascularity.*



the cases. The location of maximal intensity of the murmur was closely related to the site of drainage of the fistula. In patients with right atrial drainage the murmur was loudest at the second right intercostal space, whereas in patients with entry of the fistula into the right ventricle the maximal intensity of the murmur was in the fourth or fifth left intercostal space or at the apex.

The site of maximum intensity of the murmur was in the second left intercostal space only when the fistula was draining into the pulmonary artery or into the right ventricular outflow tract. Thus, in the majority of patients, this lesion could be distinguished from persistent ductus arteriosus by unusual location of the continuous murmur. The second sound was normally split in all patients except 2: one with pulmonary hypertension and the other with tetralogy of Fallot.

Electrocardiographic features The electrocardiogram was normal in all cases except 4. One patient who complained of dyspnoea and palpitations showed multiple supraventricular extrasystoles. In another case, diagnosed as atypical angina, the electrocardiogram showed evidence of anterolateral infarction. The patient with associated persistent ductus arteriosus showed left ventricular hypertrophy. The electrocardiogram of the patient with tetralogy of Fallot was indistinguishable from that of an uncomplicated case of tetralogy.

Chest x-rays In general the chest *x*-ray resembles that of any condition with large left-to-right shunt, except when the fistula drains into the left side of the heart, in which case the pulmonary vascularity should be normal. This was found in one of our patients with the fistula draining into the left atrium. In this patient the chest *x*-ray showed cardiomegaly with atypical signs of a much dilated left atrium and normal pulmonary vascularity (Fig. 2).

Cardiomegaly on the chest *x*-rays was closely related to the size and duration of the shunt. All 3 older patients had considerable cardiomegaly. The 65-year-old woman who was in congestive heart failure had a cardiothoracic ratio of 70 per cent, which returned to normal after operation. In the younger age-group all patients with cardiomegaly had a shunt of half the pulmonary blood flow. Right atrial enlargement was seen in 2 patients with right atrial communication.

In a third patient an unusual bulge was noted at the right atrial border, which on angiocardiographic examination was seen to be due to the much dilated coronary artery.

Dilatation of the ascending aorta was seen in only one patient.

Cardiac catheterization At cardiac catheterization the site of entry of the fistula in the right side of the heart could be detected by a rise in oxygen saturation in the particular chamber. This was true in all but 3 of our patients with a right heart communication. In 2 of these the diagnosis was confirmed at angiocardiography and in the third patient the fistula was detected at operation.

In the patient with left atrial communication, the catheter entered the left atrium through the coronary artery fistula. The left atrial pressure was 25 mmHg and the pressure curve showed both prominent a and v waves, the latter being dominant. The pressure in the pulmonary artery was moderately raised 40/0 mmHg. This may be the result of chronically raised left atrial pressure. The pulmonary artery pressure was raised to half the systemic in only one other patient who was in congestive heart failure.

The size of the shunt varied from very small to 50 per cent of the pulmonary blood flow. In 8 of the 13 patients with a detectable shunt it was less than 30 per cent of the pulmonary blood flow.

Operation Of the 11 patients who were operated on, one died during total correction of tetralogy of Fallot. Four types of operation have been performed: (1) ligation of the fistula; (2) closure of the opening of the fistula; (3) ligation and closure of the fistula; and (4) multiple mattress sutures.

In one patient the continuous murmur persisted after ligation of the fistula. In another patient, with drainage of the right coronary artery into the right ventricle, the fistula was closed by multiple mattress sutures.

During operation there was transient ST segment elevation on the electrocardiogram. In a third patient, with the left coronary artery draining into the left atrium, the opening of the fistula was closed through the left atrium. This patient developed T wave inversion in the left praecordial leads and these changes were present even 4 years after operation.

Discussion

From this study of 17 patients and from the reports, it is clear that the majority of the patients are asymptomatic clinically. Most of them are referred because of the murmur de-

tected at a routine examination. Symptoms are related to the size and duration of the shunt (Neufeld *et al.*, 1961). If the shunt is large there is a tendency for the patient to be symptomatic at an earlier age. A continuous murmur was invariably present. The quality of the murmur has not been very helpful in the diagnosis, but Gasul *et al.* (1960) reported accentuation of the diastolic component.

According to them this was a helpful sign in differentiating this lesion from persistent ductus arteriosus. While this auscultatory phenomenon was again emphasized recently by Barnes, Cheung, and Wu (1969), most authors failed to recognize it (Neufeld *et al.*, 1961; Effler *et al.*, 1967). The site of the maximum intensity of the murmur is closely related to the site of drainage (Neufeld *et al.*, 1961) and diverges from the second left intercostal space, the classical site of the continuous murmur of persistent ductus arteriosus, except where the fistula drains into the right ventricular outflow tract or pulmonary artery. Patients who presented with only a systolic murmur which later developed into a continuous murmur have been reported (Biörck and Crafoord, 1947; Davison, McCracken, and McIlveen, 1955; Barnes *et al.*, 1969).

The *electrocardiogram* of this lesion has no specific pattern. Surprisingly in most of the patients the electrocardiogram was normal. If a particular chamber was hypertrophied as a result of the shunt, this was reflected in the electrocardiogram. Rarely it shows evidence of ischaemia, as seen in one of our cases (Edwards, 1958; Sabiston *et al.*, 1963; Dedichen, Skalleberg, and Cappelen, 1966).

On the other hand, radiological examination of the chest has been helpful in the diagnosis of this lesion. The receiving chamber may be enlarged as seen in one case with left atrial and in two cases with right atrial entry.

An unusual cardiac silhouette may be seen, this being usually due to the much dilated and tortuous coronary arteries (Scott, 1948; Valdivia, Rowe, and Angevine, 1957; Swan *et al.*, 1959). There is one report of intracardiac calcification (Colbeck and Shaw, 1954) which was thought to be an aortic valve calcification while the patient was alive, but at necropsy it proved to be a calcified aneurysm of the coronary artery.

Gasul *et al.* (1960) emphasized the dilatation of the ascending aorta on the chest *x*-ray, but this was seen in only one of our patients.

Cardiac catheterization in most cases localizes the site of the shunt. As shown by the present series the shunt in this lesion can vary from very small to more than 50 per cent of the pulmonary blood flow. Right heart pressures were raised only in the presence of congestive heart failure, or where the fistula was communicating with the left atrium. However in the one reported case of a fistula draining into the left atrium, the findings at right heart catheterization were normal (Mozen, 1956).

Aortography and selective coronary arteriography are the two methods of choice in demonstrating the fistula.

In some patients, because of the huge 'run off' in the fistula, an aortic root injection may fail to opacify the normal coronary arteries. In these patients the normal coronary arteries should be demonstrated by selective coronary arteriography before operation is recommended. In the present series aortography invariably showed a much dilated and tortuous vessel, but there have been reports in which the coronary artery was not or only minimally enlarged (Biörck and Crafoord, 1947; Essenberg, 1950; Neufeld *et al.*, 1961). Aortography also clearly showed the termination of the fistula.

From this study, it is clear that patients with this lesion develop symptoms at a young age only in the presence of a large shunt. All the other patients continue to be asymptomatic until the third decade of life when they may slowly develop congestive heart failure. Once the shunt is removed they might become normal, as seen in one of our operated patients. Other complications such as bacterial endocarditis (Jacobi and Heinrich, 1933; Sanger *et al.*, 1959) or rupture of the dilated vessel (Habermann, Howard, and Johnson, 1963) are rare. Any rational management of this lesion by operation should take into account these various aspects of the natural history of the lesion. Operation should be recommended for the younger age group only if the shunt is very large; otherwise they can safely wait until they reach the second or third decade of life.

However, even in small shunts the involved vessel might become very tortuous and dilated, and might interfere with the mechanical functioning of the heart. Operation may be considered in these cases if only to alleviate this mechanical burden.

Without serial cardiac catheterization, angiocardiography, and a detailed study of coronary circulation, it is difficult to answer questions, such as, whether the lesion is progressive, how does the coronary circulation adapt itself to a 'run off' through such a fis-

tula, and what is its effect on the myocardial function and metabolism?

These are important problems, the solution to which should help to plan the management of these patients in a better way.

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